High-Flow Traumatic Carotico-Jugular Fistula Manifesting as Venous Hypertensive Encephalopathy

A Case Report

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Summary

We report the clinical and angiographic findings in a patient who presented with venous hypertensive encephalopathy secondary to a traumatic carotico-jugular fistula. Endovascular entrapment of the fistula by occluding the common carotid artery and internal jugular vein at the base of the skull resulted in near total improvement of the patient's neurological status.

Introduction

Arteriovenous fistulae occur in less than 4% of patients with trauma of the neck vessels. Most of these present with a pulsatile neck mass. Interventional radiological techniques offer an efficient and minimally invasive mode of therapy to treat these patients. High flow vascular malformations such as dural arteriovenous fistulae are known to produce elevation of venous pressures and present with features of venous hypertensive encephalopathy. We report the clinical and angiographic findings in a young man who presented with venous hypertensive encephalopathy due to a post-traumatic carotico-jugular fistula and discuss its management.

Case Report

This 35-year-old man sustained a penetrating injury on the left side of neck. This was associated with profuse haemorrhage from the wound and was treated by emergency surgery, the details of which are unavailable. Two years later, he began having left sided dull headache associated with vomiting. Over a period of ten months, the intensity of headache and frequency of vomiting had increased. This was associated with progressive confusion and memory loss, as well as neurological deficits in the form of aphasia and progressive right-sided hemiparesis, which left the patient mute and bed bound. He also had become progressively apathetic with deterioration of the level of sensorium over the week before presentation at our institute.

Physical examination revealed an ill-defined pulsatile mass on the left side of the neck, with an audible bruit on auscultation. There were no signs of cardiac decompensation. Neurological examination revealed a drowsy, but arousable patient with a Glasgow Coma Scale score of E3 M4 V1. Fundoscopy revealed bilateral papilledema. Examination of the motor system revealed increased tone and paralysis of the right upper and lower limbs.

Plain and contrast enhanced CT scans of the brain (figure 1A,B) revealed multiple, intensely enhancing, serpiginous vascular structures in both the supratentorial and infratentorial compartments. CT sections at the level of the skull base (figure 1C) revealed gross dilatation of the left internal jugular vein (IJV). There was dif-

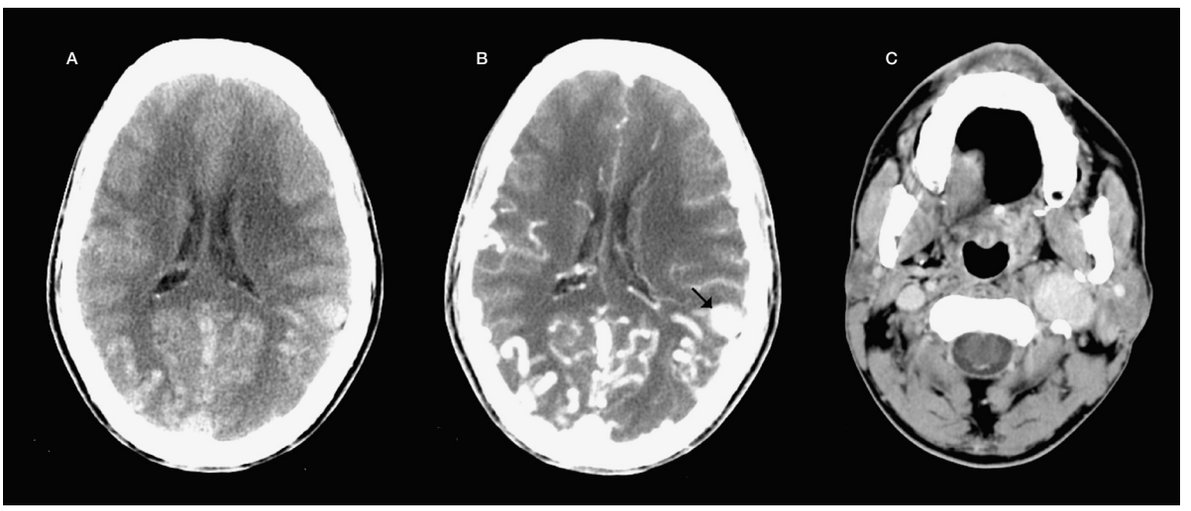

Figure 1 Plain (A) and contrast enhanced (B) CT scans at the level of the body of the lateral ventricles revealed multiple tortuous vessels - some of which showed venous ectasia (arrow). Contrast enhanced CT scan at the level of the 2nd cervical vertebra (C) revealed gross dilatation of the left internal jugular vein and non-visualization of left internal carotid artery.

fuse cerebral edema with effacement of the ventricles and cisternal spaces of the brain.

Cerebral angiography (figure 2) revealed a high-flow direct arteriovenous fistula originating from the proximal part of the left internal carotid artery (ICA), draining into a dilated and tortuous left IJV. There was no antegrade drainage from the left IJV. Instead, blood was seen to flow from the fistula, retrogradely within the IJV, to enter the left transverse sinus, torcular

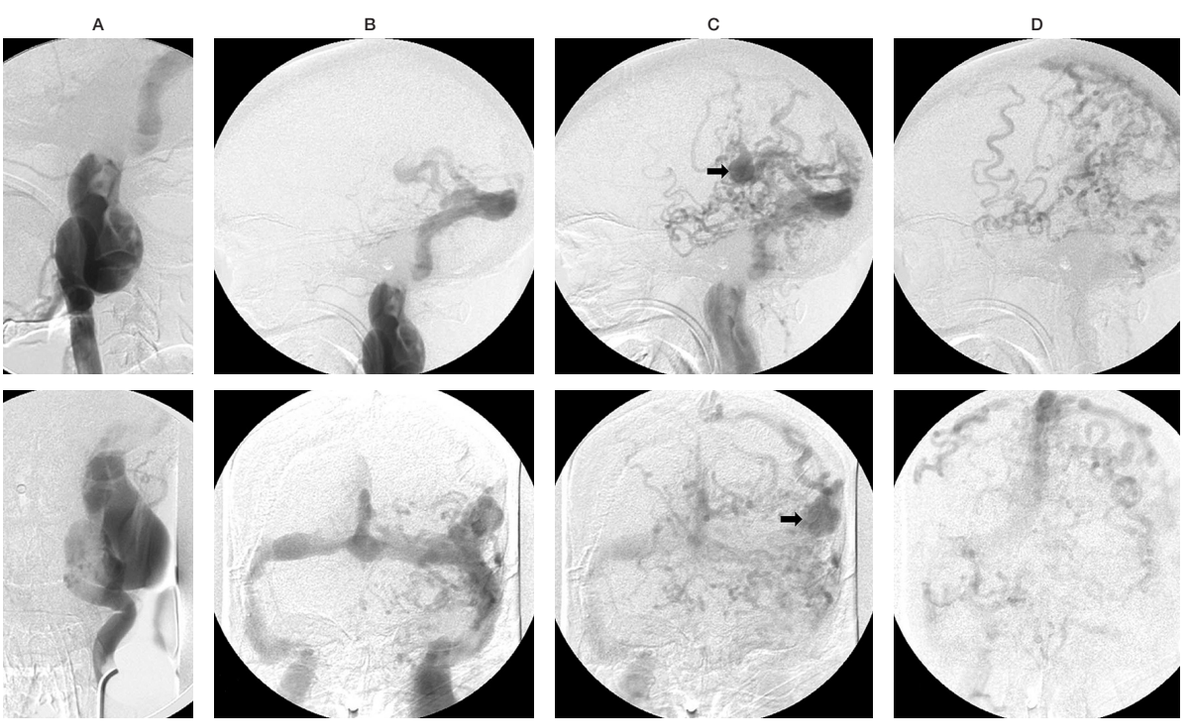

Figure 2 Serial digital subtraction angiographic images (A-D) of left common carotid artery injection in lateral (top panel) and frontal (bottom panel) revealed immediate opacification of the left internal jugular vein (A) and retrograde flow into the dural venous sinuses (B). Left hemispheric cortical veins show retrograde flow (C,D). Note the venous sac involving the vein of Labbe (arrow in C).

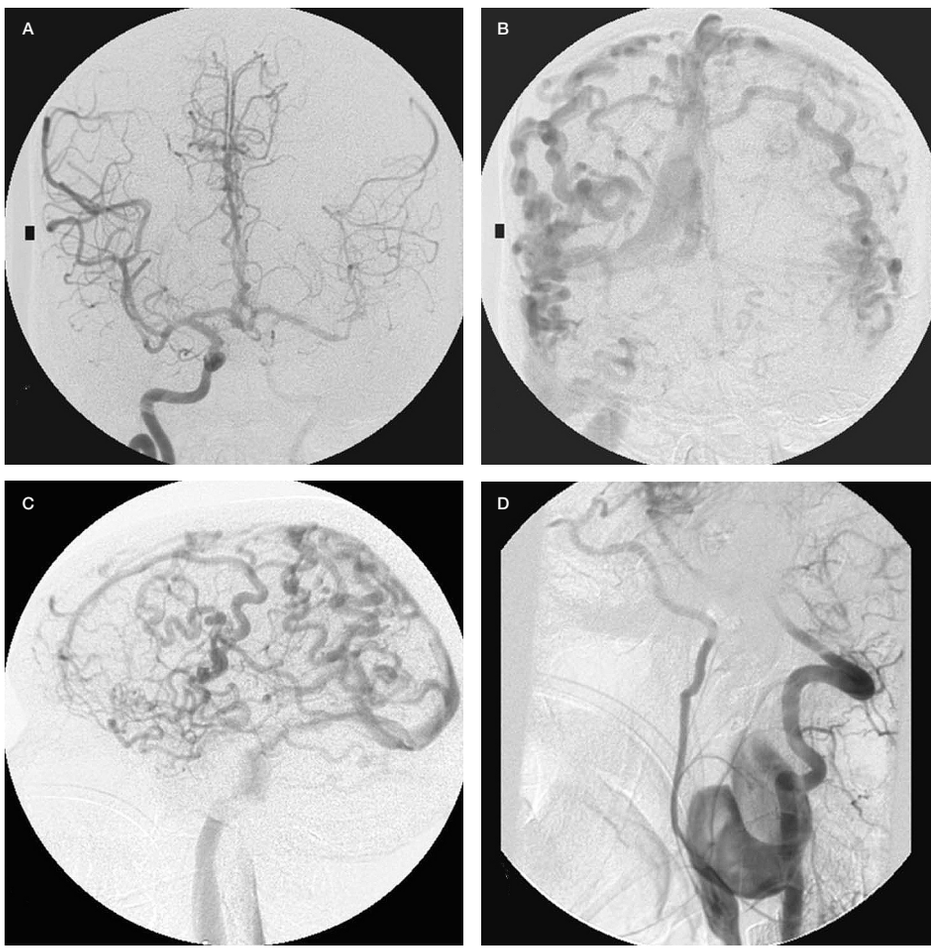

Figure 3 Digital subtraction angiogram of right common carotid artery injection in arterial (A) and late venous phases (B), revealed supply to both cerebral hemispheres from right internal carotid artery. Stasis within the tortuous and dilated veins was noted in the venous phase (19 sec after injection) (C). Opacification of the fistula on left vertebral injection (D) by retrograde flow through the distal segment of the left internal carotid artery was noted.

and straight sinus. From the straight sinus, blood was seen the flow into multiple cortical veins, that drained into the posterior third of the superior sagittal sinus (SSS). Blood from the left transverse sinus was also noted to flow into multiple cortical veins in the left temporal and occipital regions with a large venous sac in the region of the vein of Labbe. These veins were seen to drain into the posterior third of the SSS, which continued as the right transverse sinus and right IJV.

Right carotid angiogram (figure 3A,B,C) revealed supply to both cerebral hemispheres, with prolongation of venous phase up to 26 seconds. The deep venous system of both cerebral hemispheres failed to opacify on right carotid injection. Vertebral artery injection (figure 3D) revealed opacification of the fistula by retrograde flow across the posterior communicating artery and distal segment of the internal carotid artery.

Attempts at placement of a stent graft across the site of fistula were unsuccessful due to the inability to cannulate the distal segment of the ICA, owing to its small caliber and presence of the high flow fistula. In view of the high flow and the dilatation of the IJV distal to the fistula, balloon embolization of the fistula was considered to be unsafe. Occlusion of the left IJV at the level of skull base and occlusion of the left common carotid artery (CCA) proximal to fistula was considered to be the best mode of treatment. The flow from the site of fistula into the intracranial venous system was rapid and accidental migration of the detachable balloon used for IJV occlusion was of grave concern. It was decided to protect this balloon proximal to its site of detachment, using Guglielmi Detachable Coils (GDC) through a contralateral jugular venous approach.

The carotid-jugular occlusion procedure was performed under general anesthesia. Bilateral

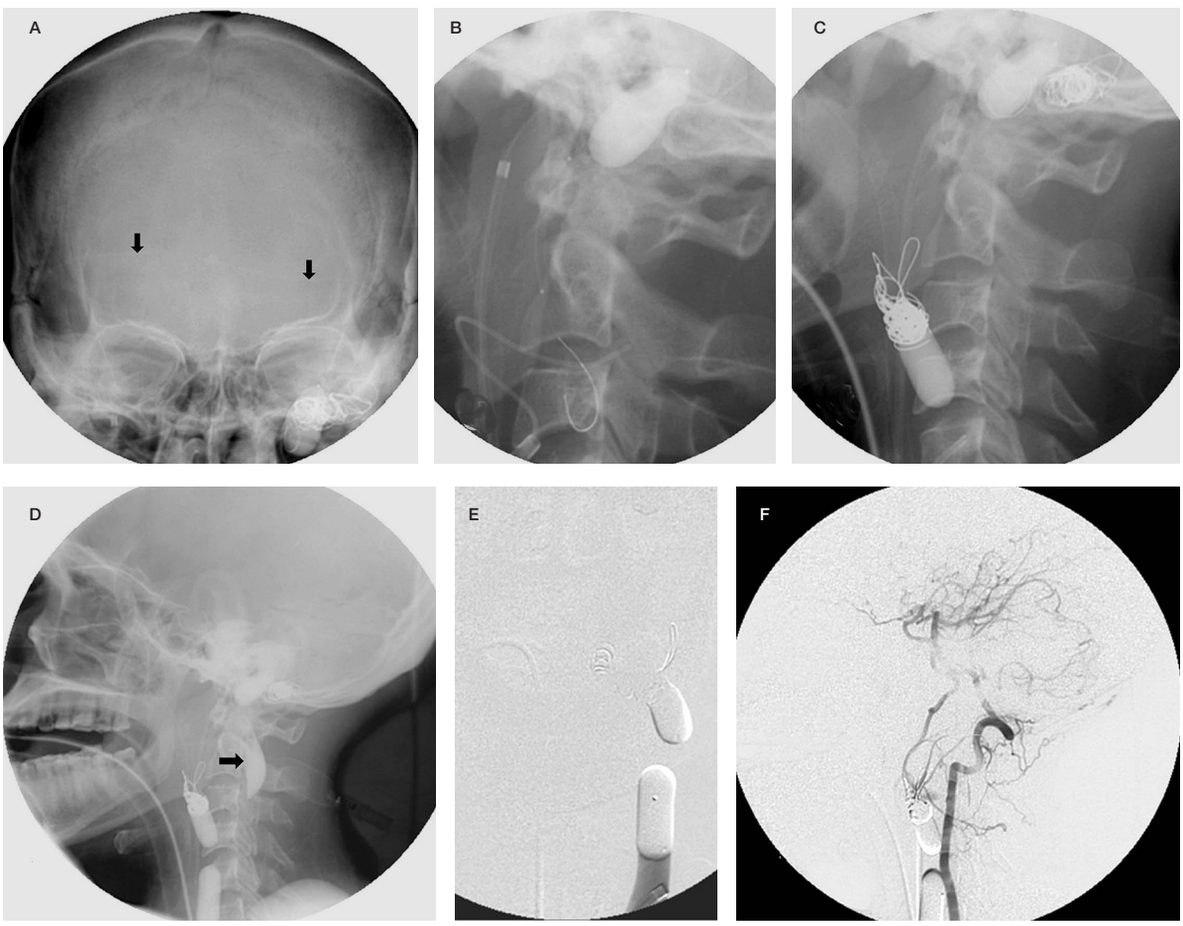

Figure 4 A microcatheter (Excel-14, arrows in A) is seen passing from right jugular access, across the torcular into the left transverse sinus and left jugular bulb. Gold valve balloon (GVB - 9) was introduced through the left common carotid artery and fistula and navigated to the left jugular bulb (B). Deploying GDC coils within the jugular bulb and distal sigmoid sinus, using the contralateral internal jugular vein access, ensured protection against intracranial migration of the balloon (C). Balloon and coil occlusion of the left common carotid artery resulted in stasis of contrast medium within the venous sac (D). Check angiogram revealed non-opacification of the fistula on common carotid injection (E), but residual flow was seen on left vertebral injection (F).

femoral arterial access and right femoral venous access were secured. An 8 French guide catheter (Vista Brite tip, Cordis Corporation, Miami, Fl.) was placed in the left CCA. A wedge pressure balloon catheter (Berman angiographic balloon catheter, Arrow International, Reading, PA) was placed in the proximal left CCA, and was inflated to achieve flow reduction across the fistula.

A 6 French guide catheter (Envoy, Cook Corporation, USA) was used to cannulate the right internal jugular vein, and a microcatheter (Excel-14, Target Therapeutics, Fremont, CA) was negotiated into the right transverse sinus, and across the torcular into the left jugular bulb (figure 4A). Through this microcatheter, a 20mm x 30cm Guglielmi Detachable Coil (GDC-18, Target Therapeutics, Fremont, CA) was deployed within the left jugular bulb and proximal IJV. A detachable balloon (GVB-9, Nycomed, Paris, France) was mounted on a 2F / 3F coaxial catheter system (Nycomed, Paris, France) and introduced into the left CCA through the 8F guide catheter. Partial inflation of the balloon carried it across the fistula, into the IJV. The balloon was positioned in the left jugular bulb and inflated (figure 4B).

The microcatheter that was positioned at the jugular bulb through the venous access was withdrawn a little and the rest of the GDC coil

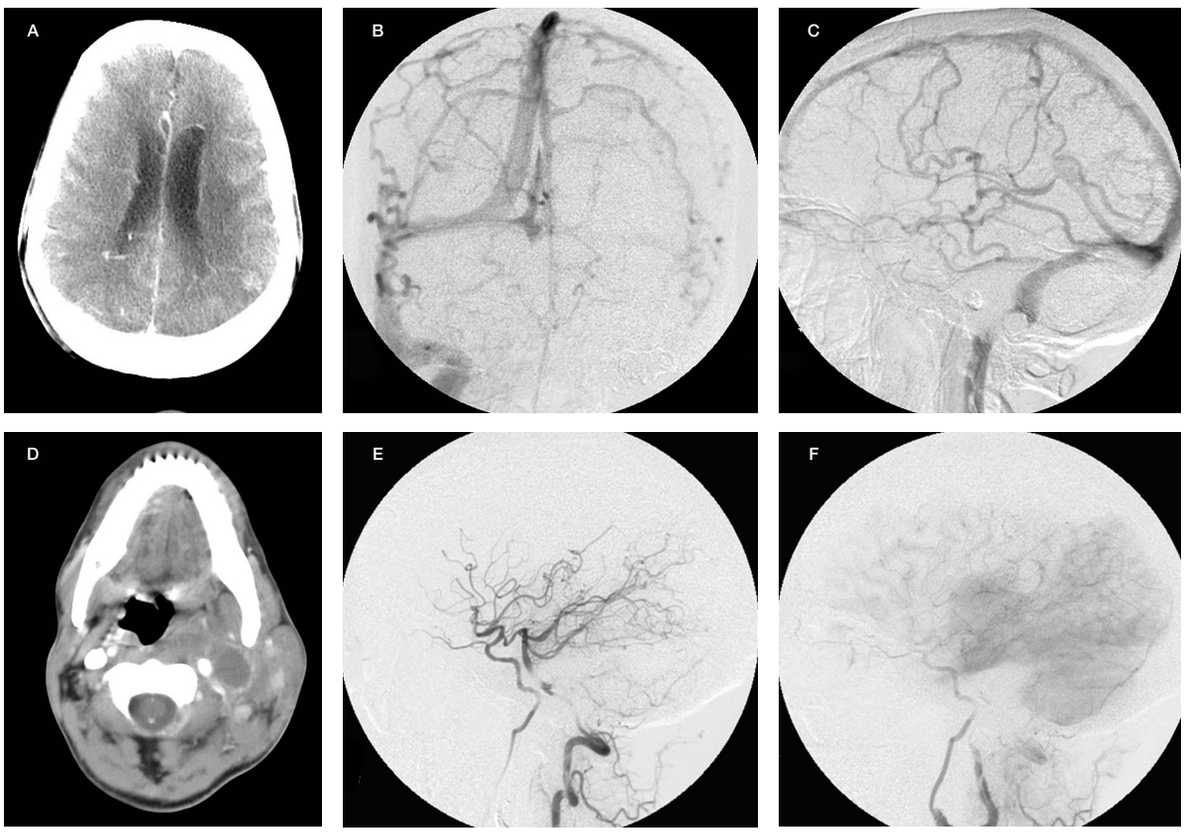

Figure 5 Contrast enhanced CT scans of the brain one month after the procedure revealed disappearance of the intracranial serpiginous veins (A). The venous sac at the vein of Labbe has also shown reduction in size. Venous phase of cerebral angiogram in frontal (B) and lateral (C) projections revealed normal caliber of intracranial veins. Contrast enhanced CT scan the level of the 2nd cervical vertebra revealed thrombosis of the jugular venous sac (D). Left vertebral angiogram (E and F) revealed persistent opacification of the fistula by retrograde flow. Flow within the fistula was extremely slow.

was deployed within the jugular bulb and distal part of sigmoid sinus, superior to the balloon; thus protecting it against migration (figure 4C). Subsequently, the left CCA was occluded by multiple fibered coils and two detachable balloons (GVB-9 and GVB-12, Nycomed, Paris, France). Post-procedure left carotid angiogram revealed complete occlusion of the left CCA and non-opacification of the fistula. Right carotid angiogram revealed stasis within the cortical veins. Minimal residual flow into the fistula through the posterior communicating and distal internal carotid artery was noted on vertebral artery injection, but there was complete stasis of contrast medium within the venous sac (figure 4D,E,F).

During the post-procedural period, the patient was treated with intravenous injection of 20% mannitol, dexamethasone, aspirin and ticlopidine. Continuous intravenous infusion of heparin was continued for three days after the procedure. Within 12 hours, the level of sensorium improved to a GCS of E3 M6 V1. He had a persistent global aphasia and right hemiparesis. With rehabilitative therapy and speech therapy, the limb power and speech improved over the next week. The patient was discharged 25 days after the procedure, with mild weakness of right upper and lower limbs.

One month later, the patient was readmitted with complaints of painful neck swelling. Examination revealed a firm and tender swelling on the left side of the neck, which approximated the location and size of the venous sac. There were multiple palpable, tender cervical lymph nodes. These subsided on therapy with broad-spectrum antibiotics and anti-inflammatory medication. Angiographic study (figure 5)

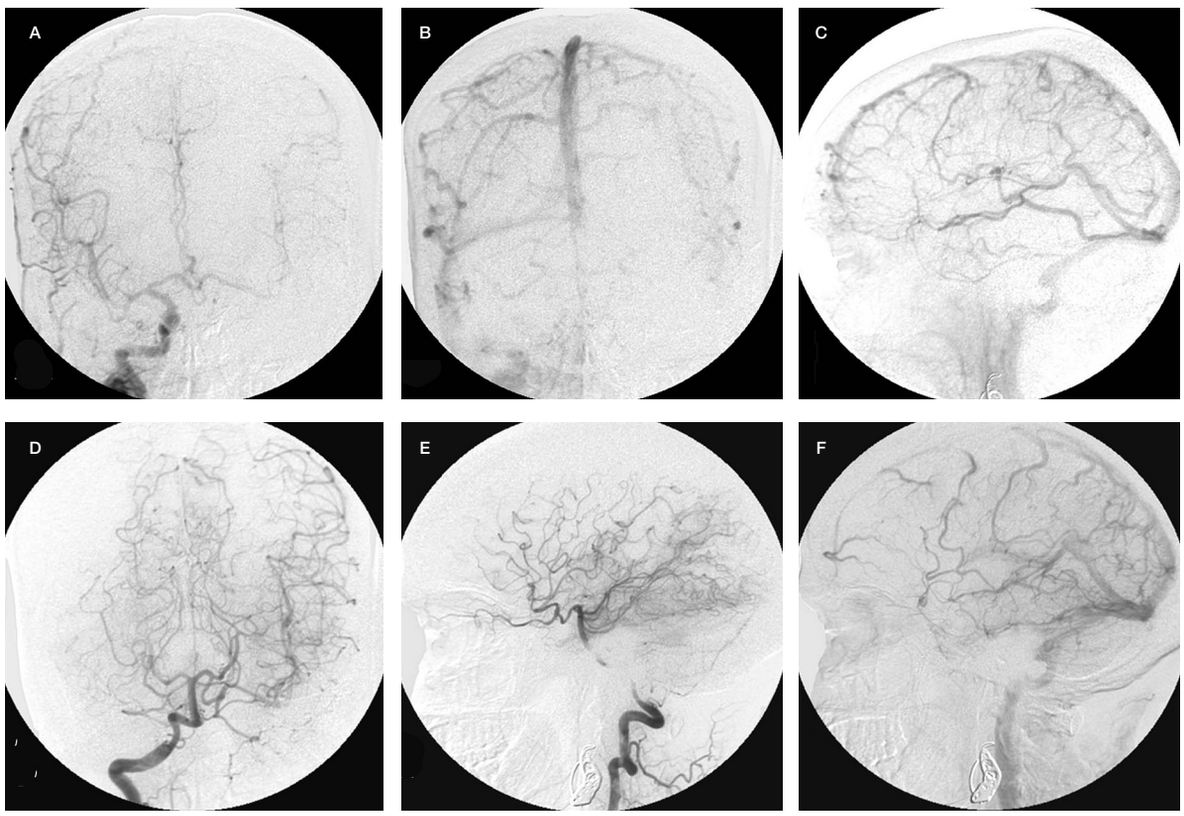

Figure 6 Follow up angiogram two years after the procedure revealed normal caliber of intracranial veins (A to C). Left cerebral hemisphere was seen to derive supply from both right carotid and posterior circulation (A and D). Complete obliteration of the fistula was confirmed by absence of flow in late venous phase of vertebral angiogram (F).

revealed persistence of filling of the fistula from the vertebrobasilar circulation, by retrograde flow through the distal internal carotid artery, via the posterior communicating artery. The size of venous sac had decreased and there was no flow into the intracranial venous system. Multiple small veins were seen to drain the residual venous sac.

On right carotid angiogram the cortical veins were seen to be of normal caliber and angiographic circulation time was normal. Embolization of the residual fistula by retrograde approach through the posterior communicating artery was attempted. However, the microcatheter could not be negotiated through the posterior communicating artery into the ophthalmic segment of the ICA due to an acute angle between the two arteries.

The patient has been on regular clinical follow up since then. Two years after the procedure, he has mild residual right-sided facial weakness and mild weakness of right upper limb. He has returned to his work. Follow-up angiogram, two years after the procedure (figure 6) revealed total closure of the fistula. There was no filling of the distal segment of the ICA and venous sac from vertebrobasilar circulation. Left external carotid artery was seen to opacify via multiple collaterals between left vertebral artery and the occipital artery.

Discussion

Arteriovenous fistulae account for less than 4% of complications following vascular trauma in the head and neck region [1]. While penetrating trauma due to war-time injuries is the most common cause [2], there have been reports of occurrence of carotico-jugular fistulae after neck surgery and infection [3] and even as iatrogenic complication of internal jugular vein cannulation [4]. Congenital carotico-jugular fistulae are rare and usually appear early in life.

Carotico-jugular fistulae usually present as

pulsatile neck masses. The high flow arteriovenous shunt at the fistula manifests as tinnitus, arterial steal and as high output cardiac failure. Lower cranial nerve palsies, dysphagia and stridor can also occur as a result of mass effect due to the dilated venous components. Extracranial arteriovenous fistulae have rarely been reported to cause progressive neurological deficits. Horiuchi et Al have found only three cases of external carotid-jugular fistulae, in addition to their case, which presented with features of posterior circulation insufficiency [5].

The patient described in this report presented with progressive cerebral dysfunction that has been described to occur with cerebral venous hypertension. In 1998, Hurst et Al [6] have described the clinical and radiological findings in five patients who had venous hypertensive encephalopathy secondary to dural arteriovenous fistulae. The mode of presentation in these patients was with headache, worsening confusion, disorientation, focal neurological deficits and dementia; progressing to a state of inability to care for themselves. Cognitive dysfunction predated the onset of any focal neurological deficits.

The insidious onset and temporal profile of progression of symptoms in our patient were similar. The headache and vomiting were initial manifestations of increase in intracranial pressure. The retrograde transmission of pressure to the left hemispheric cortical veins such as vein of Labbe was responsible for early presentation with aphasia, confusion and right hemiparesis. The deterioration of level of sensorium at the time of presentation could have been secondary to diffuse cerebral dysfunction or due to brainstem congestion. The rapid improvement in level of sensorium and progressive improvement of speech as well as the motor function after balloon embolization confirms the causal relationship between the cerebral venous hypertension and the clinical findings. Zeidman et Al [7] reported progressive improvement of cognitive function and motor deficits, as well as reversibility of white matter changes after treatment of dural arteriovenous fistulae.

To the best of our knowledge, the occurrence of cerebral venous hypertension causing diffuse encephalopathy in a case of traumatic extracranial arteriovenous fistula has not been described in literature. The high flow across the fistula and absence of antegrade venous drainage resulted in retrograde flow into the internal jugular vein and dural venous sinuses. The lack of antegrade drainage in this patient could have been a result of the prior surgery that was performed to arrest the bleeding when the patient sustained the penetrating injury.

CT scan of the brain, performed prior to therapy revealed diffuse cerebral edema and the presence of multiple serpiginous vascular structures. Angiography confirmed these structures to be dilated and tortuous cortical veins forming a superficial network to redistribute the elevated cerebral venous pressure. The presence of serpiginous engorged veins in the venous phase of cerebral circulation has been termed as the "pseudophlebitic pattern". This finding has been described to occur in vein of Galen malformations, brain arteriovenous malformations and dural arteriovenous malformations with cortical venous drainage [7]. To the best of our knowledge, the association of the pseudophlebitic pattern with extracranial arteriovenous fistulae has not been described in literature.

Therapeutic options in these patients include surgical repair or ligation and interventional neuroradiological therapy. Surgical ligation involves extensive dissection of scarred tissue, which increases the complexity of surgery on a high flow arteriovenous fistula. This was considered to be difficult in our patient. Endovascular techniques are an effective and elegant alternative to surgery. The optimal technique for management of this lesion was to deploy a stent graft to occlude the fistula while preserving the patency of the artery. This was not possible in our patient due to the failure to access the lumen of the distal segment of the internal carotid artery, owing to the small caliber and presence of the high flow fistula. Stent grafts have been used in cases of cervical internal carotid artery aneurysms as well as peripheral arteriovenous fistulae, with good results. Embolization with an accurately positioned detachable balloon is equally effective in occluding the fistula [8].

The large caliber and high flow within vascular structures permits easy catheterization of the site of fistula. In the present case, the presence of a wide arteriovenous communication and presence of a hugely dilated internal jugular vein made carotid occlusion unavoidable. Inflation of an angiographic balloon catheter in the proximal common carotid artery permitted flow reduction, to prevent migration of the de-

tachable balloons and coils into the intracranial venous system. Similarly, the detachable balloon that was used to occlude the jugular bulb was protected from migration by detaching long GDC coils on both sides of the balloon. Though there was residual flow in the fistula even after occlusion of the IJV and CCA, thrombosis of the venous sac resulted in progressive obliteration of the fistula on follow up angiographic studies.

As has been demonstrated in other conditions such as vein of Galen malformations and dural arteriovenous fistulae associated with severe venous hypertension, cessation of flow across the fistula results in rapid and complete resolution of symptoms. Endovascular techniques offer an easy and effective solution in management of such lesions.

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